Frontotemporal Dementia

American Academy of Neurology

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Frontotemporal Dementia

- **Definition: clinicopathologic condition consisting of deterioration of personality and cognition assoc. with prominent frontal and temporal lobe atrophy**
- **Accounts for up to 3-20% of dementias**
  - **Third behind AD and Lewy Body Dementia in neurodegenerative dementing illnesses**

Prevalence

- **Mean age of onset 52.8 (Ratnavalli et al. Neurology 2002;58:1615-1621)**
- **Male preponderance 14:3 in one study and M=F in others**
- **Dementia prevalence of 81 per 100,000 (95% CI, 62.8 to 104.5) in the 45-64 year age group**
- **Prevalence of AD and FTD in 45-64 age group same at 15 per 100,000 (8.4-27.0)**

Frontotemporal Dementia

- **Thus, common cause of dementia in younger population**
- **Greater caregiver burden and increased dependency and health care costs**

Frontotemporal Dementia

- **Established clinical consensus criteria (The Lund and Manchester Groups, J Neurol Neurosurg Psychiatry 1994;57:416-418; Neary et. al, Neurology 1998;51:1546-1554):**
- **Core features**
  - **Insidious onset and slow progression**
  - **Early decline of**
    - **Social interpersonal conduct**
    - **Regulation of personal conduct**
    - **Insight**
  - **Early emotional blunting**

Frontotemporal Dementia

- **Supportive features:**
  - **Decline in personal hygiene and grooming**
  - **Mental rigidity and inflexibility**
  - **Distractibility and impersistence**
  - **Hyperorality**
  - **Perseverative behavior**
  - **Speech and language**

Frontotemporal Dementia

- **Neuropsychology:**
  - **Impaired frontal lobe tests in absence of severe amnesia, aphasia, or visuospatial deficits**
- **Imaging:**
  - **Atrophy or decreased uptake in the frontal or anterior temporal lobes (bilateral or unilateral) by MRI, CT, PET, SPECT (The Lund and Manchester Groups, J Neurol Neurosurg Psychiatry 1994;57:416-418; Neary et. al, Neurology 1998;51:1546-1554)**

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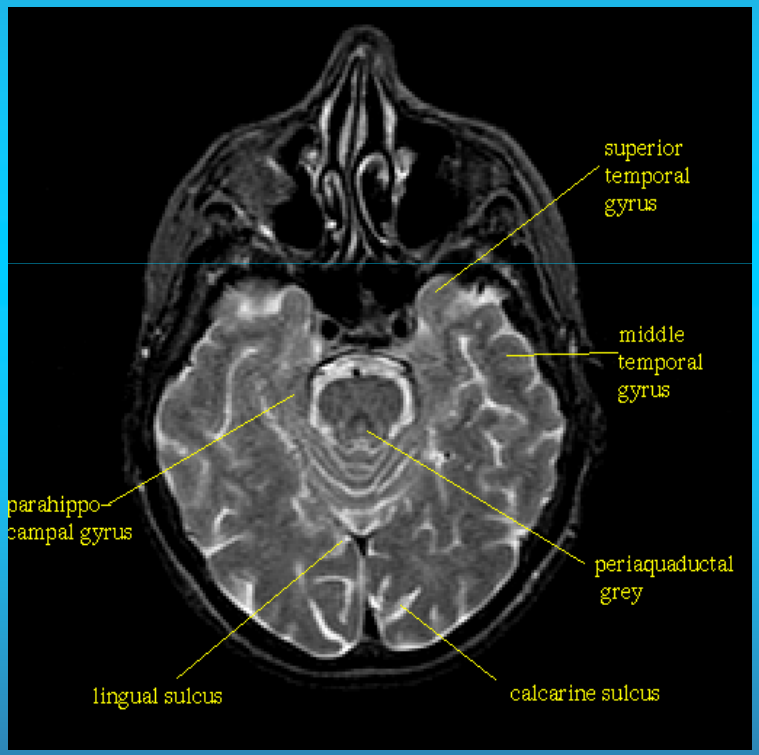

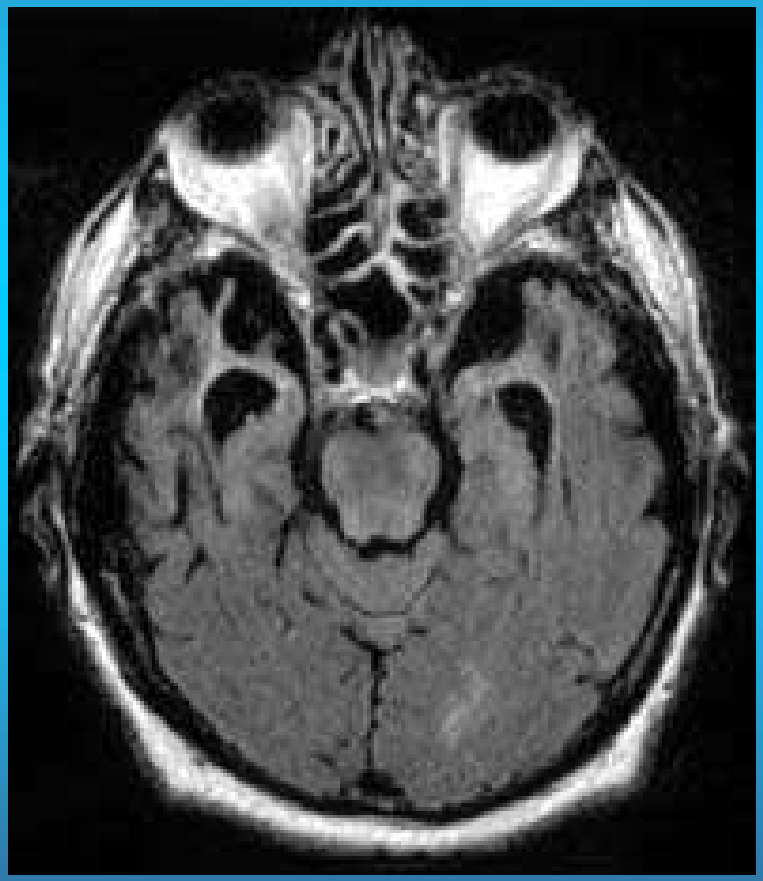

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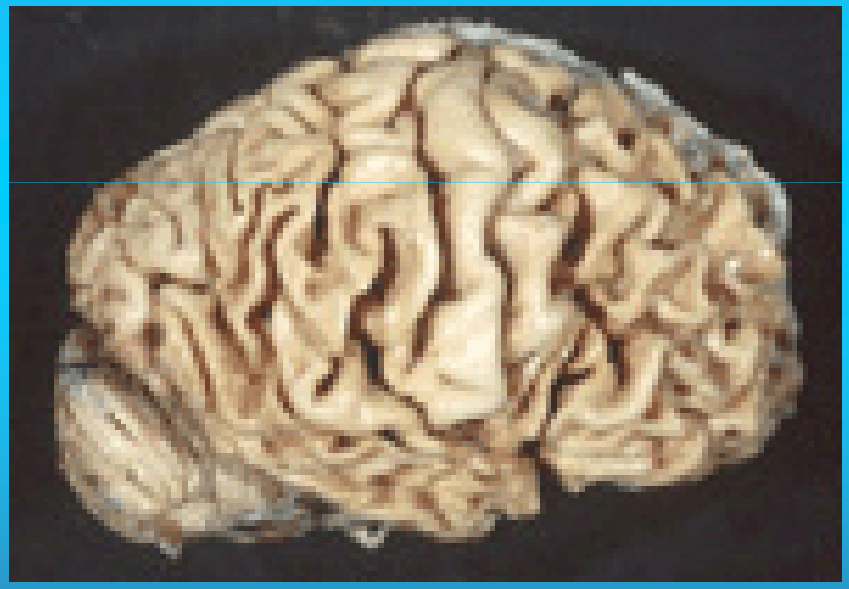

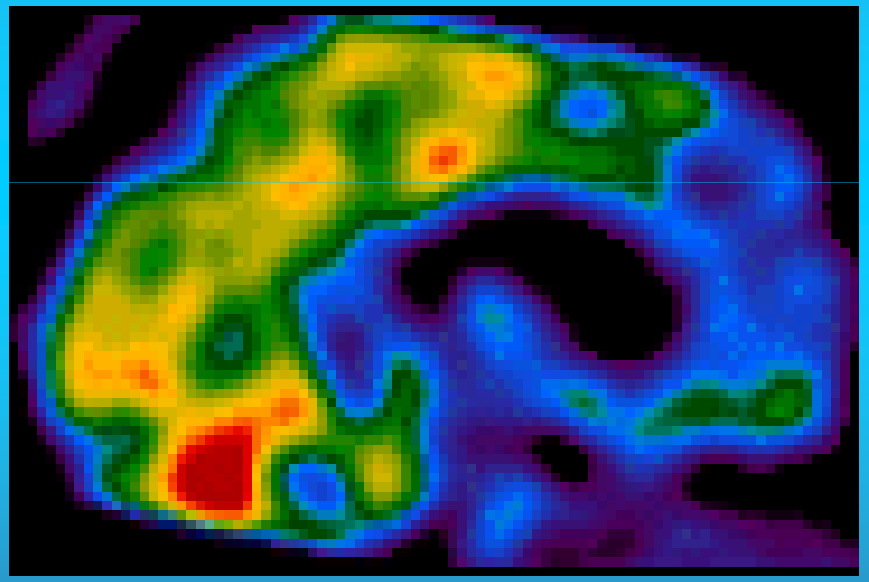

Frontotemporal Dementia

- **Recent study showed of 42 FTD cases, 19 had at least 1 other family member affected (Chow et al., Arch Neurol. 1999;56:817-822)**
  - **1/3 had a positive family history in this study, but others much lower**
- **Appears to be inherited in autosomal dominant fashion in those patients**
- **Linked to chromosome 17 (tau gene) and chromosome 3**
- **Association with motor neuron disease (ALS)**

Pathology

- **Prominent frontal and temporal lobar atrophy**
- **Atrophy may be associated with Pick's bodies, tauopathy, nonspecific superficial cortical neuron loss (DLDH)**

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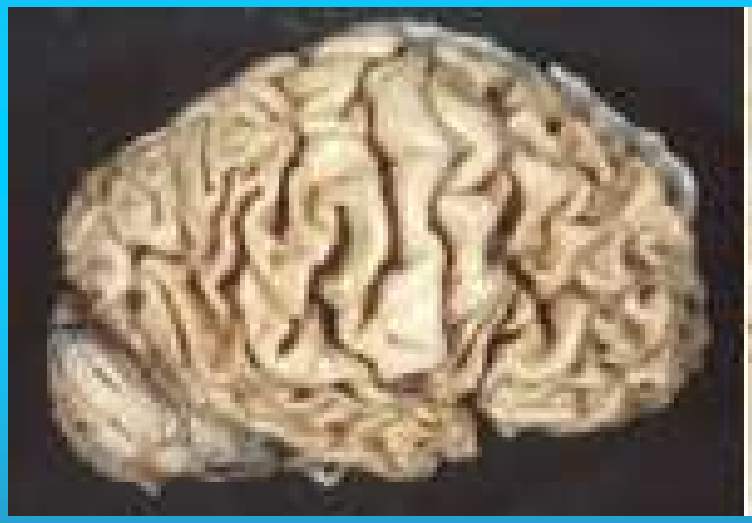

Frontotemporal Dementia

- **Term is conventionally used in clinical and pathological diagnoses in standard clinical setting**
- **No other specific code comes reasonably close to capturing disease**
- **Dementia or AD diagnosis (290 or 331) ignores established diagnostic criteria and allows no tracking of FTD**

Frontotemporal Dementia

- **Differs from the other codes including AD, frontal dementia, Pick's disease**
  - **Use of these codes inappropriate as not capture the age of onset, duration of illness, genetic factors, and impact on caregiver, society, and economics**
- **AD → older, different duration, less clear genetics**
- **Frontal dementia → no temporal lobe involvement, genetics differ**
- **Pick's disease → not capture spectrum of FTD**

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- **Implications for FTD different from other dementias/AD:**
  - **Greater caregiver burden and increased dependency and health care costs**
  - **Patients see codes and think they have some other disease**

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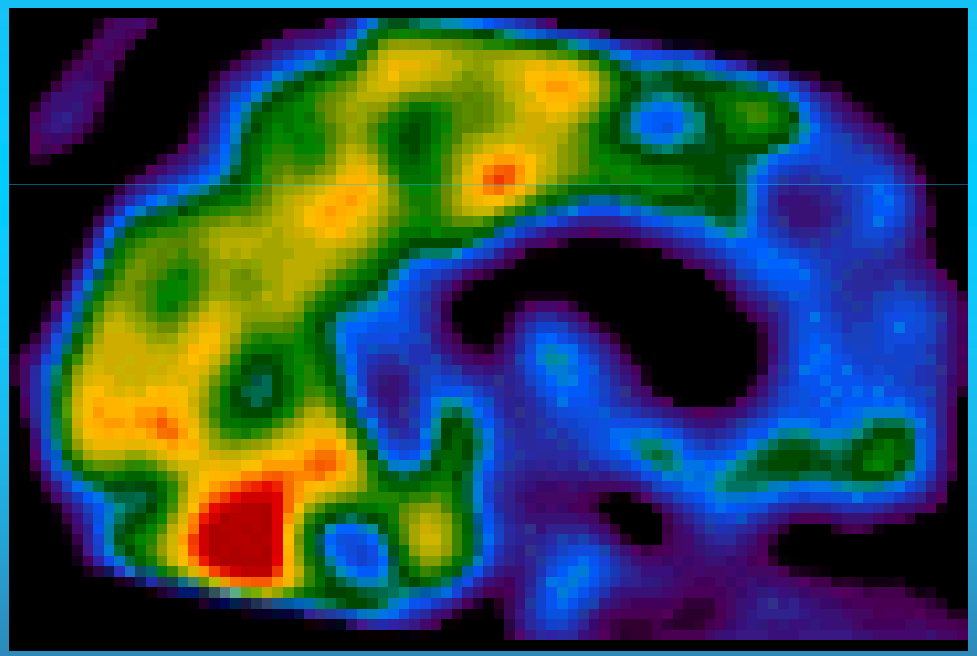

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